Adamantinoma of the Tibia Masked by Fibrous Dysplasia

Report of Three Cases

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Three cases of adamantinoma of the tibia associated with and mimicking fibrous dysplasia or osteofibrous dysplasia are reported in children aged three, nine, and 16 years. The roentgenographic features were typical of intracortical fibrous dysplasia (osteofibrous dysplasia). These entities are not two distinct diseases, but rather are different histologic expressions of essentially the same process, which varies depending on location (predominantly intracortical or medullary) and age. Adamantinoma in children under ten years of age is not as rare as reported in the literature and was found in two of 14 cases in our files (14.3%). In some cases the fibrous dysplasialike component predominates over the scarce epithelioid islands of tumor cells and consequently is not recognized as adamantinoma. That may explain the frequent recurrences after incomplete excisions of supposed intracortical fibrous dysplasia lesions in young children. An extensive histopathologic study of the biopsy and/or surgical specimen by a specialized pathologist is therefore advisable.

Adamantinoma is an uncommon but well-established neoplasm that occurs predominantly in the tibia and rarely in other long bones. In exceptional cases the lesion is found in short bones. Moon and Mori[16] carefully reviewed the literature and found 195 well-documented cases, to which they added five new cases. Only six cases (3%) were in children under the age of ten years. Moon and Mori[16] prefer the name "adamantinoma of the appendicular skeleton." The main clinical and roentgenographic features are described in that and several other reports, including some larger series.[3,10,24,25] The latter authors described four basic histologic patterns: basaloid, spindled, squamoid, and tubular, or a combination of these. The basaloid pattern is similar to basal cell carcinoma or to the cutaneous adnexial tumors, particularly the sweat gland carcinoma.

The great variety of histologic patterns and the tumor's predominant tibial location are the principal reasons for the controversy over the pathogenesis of the tumor cells, especially their epithelial *versus* vascular (endothelial) nature.[5,10,14] At present, the ultrastructure[1,8,17–20,22,23,26] and the more recently described immunohistochemical properties[9,17,18,21] favor an epithelial origin of this neoplasm. There is an association of fibrous dysplasia with adamantinoma of the tibia.[2,3,6,7,12,15,24] Weiss and Dorfman[25] interpreted the fibrous dysplasia-like areas in adamantinoma (found in seven of nine cases) as part of the spectrum of mesenchymal differentiation of which the tumor is capable.

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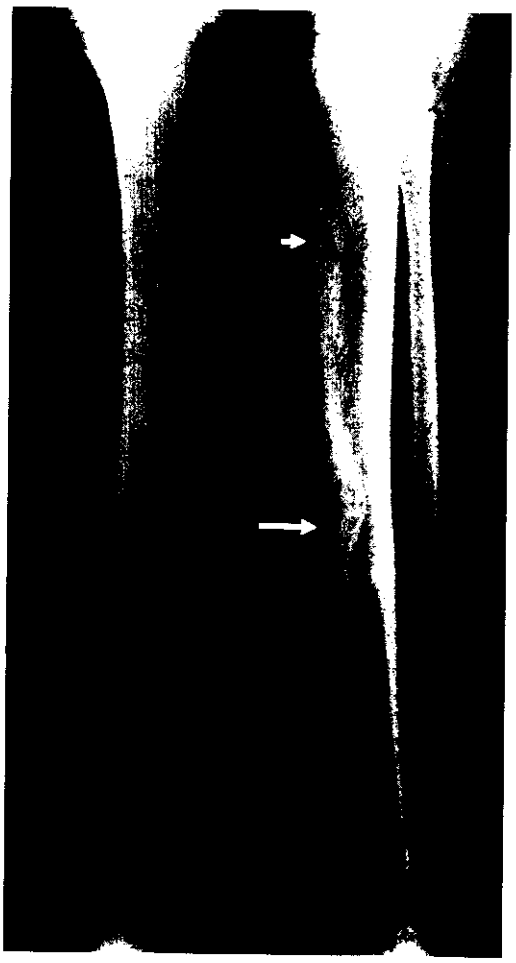


FIG. 1. Case 1. Anteroposterior (AP) and lateral roentgenograms of the tibial midshaft of a nine-year-old girl. There is a lucent multilocular lesion with a sclerotic border on the anterior aspect of the midshaft, primarily involving the cortex (large arrow). A similar but smaller radiolucent lesion is seen on the upper one-third of the tibia (short arrow).

The roentgenographic appearance of adamantinoma, showing radiolucent solitary or multiple lesions along various lengths of the affected bone with an admixture of radiodense areas, is highly characteristic. It is very similar to that found in cases of intracortical location of fibrous dysplasia (classified by some authors as osteofibrous dysplasia[4] or ossifying fibroma[13]), generally affecting children under the age of ten years. Adamantinoma, however, is very rare in children less than ten years of age.[11,16] In these adamantinoma cases, the epithelioid tissue within a prominent stromal component is often very scarce, and a mistaken diagnosis of fibrous dysplasia or osteofibrous dysplasia is frequently made.

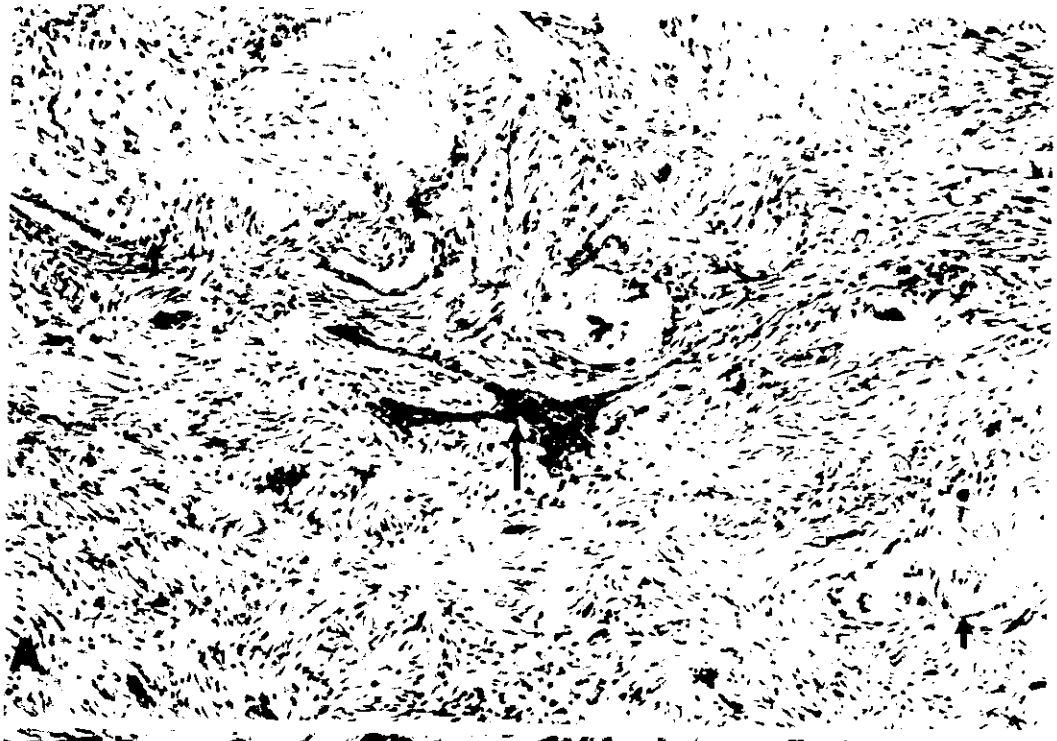
Three cases of adamantinoma of the tibia mimicking fibrous dysplasia in an intracortical location are reported. Two of the affected children were under the age of ten years; all three of the cases had similar roentgenographic features, and in each case the original pathologic diagnosis was that of fibrous dysplasia or osteofibrous dysplasia.

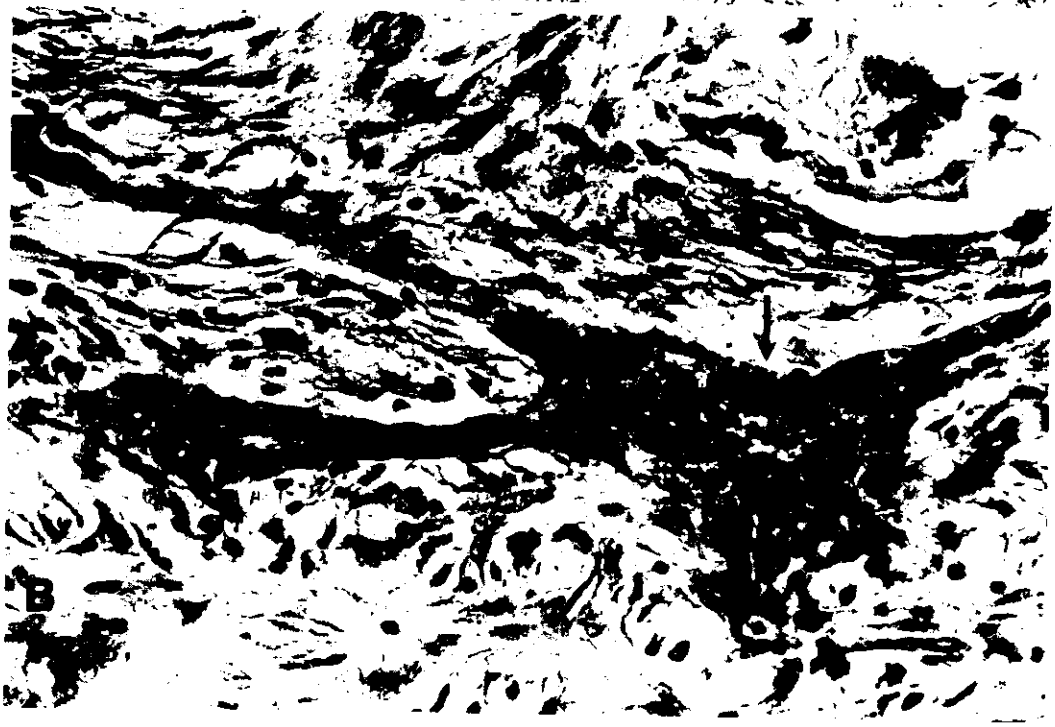
MATERIALS AND METHODS

The three cases were observed in a period of two years during 1984 and 1985, and the diagnosis of the first pathologist was fibrous dysplasia or osteofibrous dysplasia. Each diagnosis was later modified to adamantinoma of the tibia. Clinical and roentgenological information were available for the three cases. The histologic sections in two cases and the gross specimen in the third case were also available. The histologic sections of the material were examined after decalcification, embedding in paraffin, and staining by hematoxylin and eosin and Mallory's trichrome. Wilder's method was used to stain for reticulin fibers.

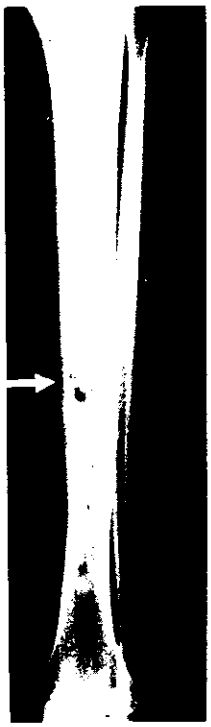
CASE REPORTS

Case 1. A nine-year-old girl complained of swelling and pain on the midshaft of the right tibia for several months. Roentgenograms showed a lucent lesion at the anterior aspect of the cortex of the tibial diaphysis, which was surrounded by reactive bone. A similar but much smaller radiolucent lesion was found at the upper portion of the tibia (Fig. 1). An open biopsy, followed by local resection of the lesions, was performed. Only small islands of tumor cells with a basaloid pattern were found. Nevertheless, histologic study of the biopsy and resection specimens confirmed the diagnosis of adamantinoma associated with active or intracortical fibrous dysplasia (osteofibrous dysplasia) with some xanthomatous zones in the fibrous stroma (Figs. 2A and 2B). The upper lesion showed only fibrous dysplasia-like lesions surrounded by bone sclerosis. The patient was reoperated upon after two years in order to correct the bowing deformity of the tibia and a recurrence





FIGS. 2A AND 2B. Case 1. Photomicrographs show some isolated tumor islands (arrows). (A) The lesion has a basaloid pattern within a fibrous dysplasialike lesion, consisting of a loose fibrous stroma with a storiform pattern and newly formed bone trabeculae (short arrows) with rims of osteoblasts on their surface (hematoxylin and eosin; original magnification, ×100). (B) The same tumor islands (arrow) at a larger magnification (hematoxylin and eosin; original magnification, ×400).



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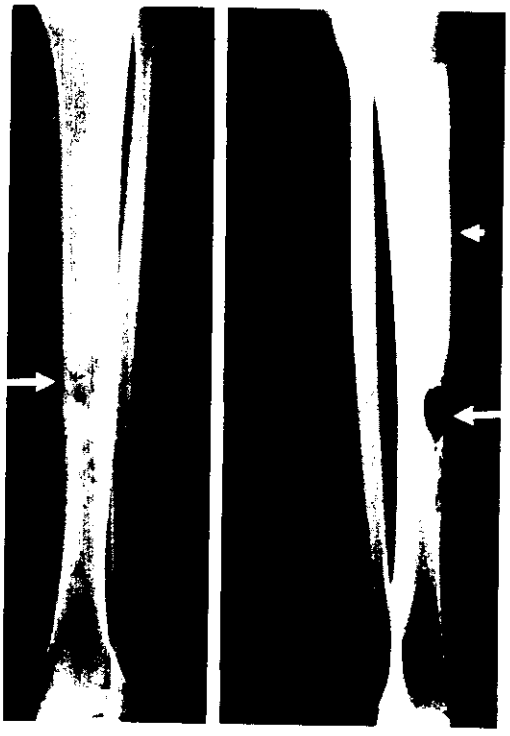


FIG. 3. Case 2. AP and lateral roentgenograms showing a larger and some smaller multifocal lucent lesions involving the anterior aspect of the tibial middiaphysis. The cortex above the lytic lesion shows evident sclerosis and a small radiolucent lesion at the upper one-third of the tibia (arrowhead).

of the upper lytic lesion. A marginal excision of this lesion and of the anterior convex deformity plus an osteotomy were performed. In this area after extensive search only one isolated small epithelioid focus was found surrounded by abundant fibrous stroma. The patient had no symptoms six months later.

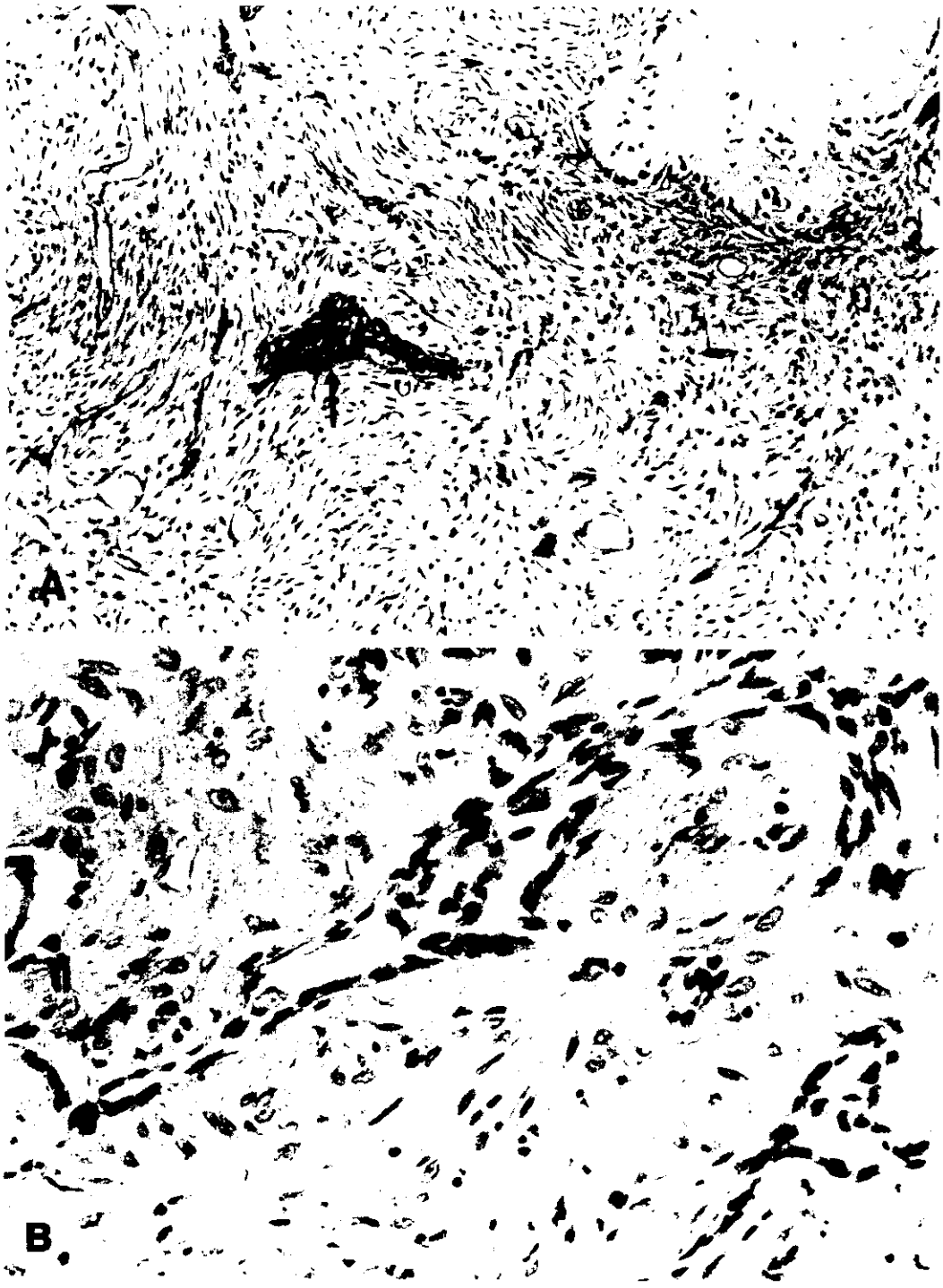
Case 2. A 16-year-old boy complained of pain in his left tibia of three month duration. Roentgenograms (Fig. 3) showed a large radiolucent, multifocal lesion and some smaller lesions involving the midshaft of the cortex of the tibia. An open biopsy and curettage were performed. Two local pathologists made the diagnosis of fibrous dysplasia and a third, adamantinoma. The diagnosis of adamantinoma associated with fibrous dysplasia was confirmed. The abundant fibrous stroma had a storiform pattern with formation of woven bone, with only a few trabeculae showing any evidence of osteoblastic rimming. Only a few solid islands of cuboidal or polyhedrical tumor cells of epithelioid appearance (Fig. 4A) and some larger spaces surrounded by cuboidal or flattened cells (similar to prominent endothelial cells) were found (Fig. 4B). This last pattern closely simulates a vascular neoplasm[5,10,14] and is not infrequent in adamantinoma. A wide resection of the anterior part of the involved tibia diaphysis was performed. The patient was well three years after the first operation.

Case 3. A three-year-old boy presented with a lucent multifocal lesion on the anterior cortex of the tibial shaft (Fig. 5). One histologic slide and the roentgenogram were submitted for review. The local pathologist and one consultant specializing in bone pathology made the diagnosis of osteofibrous dysplasia. A third bone pathologist made the diagnosis of adamantinoma associated with osteofibrous dysplasia (intracortical fibrous dysplasia; Figs. 6A and 6B), which was confirmed. The patient consulted another orthopedic surgeon and was lost to follow-up evaluation.

DISCUSSION

The coexistence of fibrous dysplasialike lesions and adamantinoma of the tibia has been reported for more than three decades,[2,3,6,7,12,15] but it seems to be more frequent than formerly believed. It is more commonly found in young children below the age of ten years. These lesions have been called ossifying fibroma by Kempson[13] and osteofibrous dysplasia by Campanacci and Laus.[4]

FIGS. 4A AND 4B. (A) Photomicrograph of Case 2 shows a solitary island of epithelioid tumor cells within a prominent fibrous stroma rich in capillary vessels and with a storiform pattern. At the upper right corner is a newly formed woven bone trabecula rimmed by active osteoblasts (hematoxylin and eosin; original magnification, ×100). (B) The same case at higher magnification shows a tubular pattern with elongated spaces devoid of blood cells and lined by cuboidal or flattened epithelioid cells. This pattern closely simulates vascular spaces, sometimes resulting in the interpretation of this lesion as tumor of vascular (endothelial) origin (hematoxylin and eosin; original magnification, ×400).



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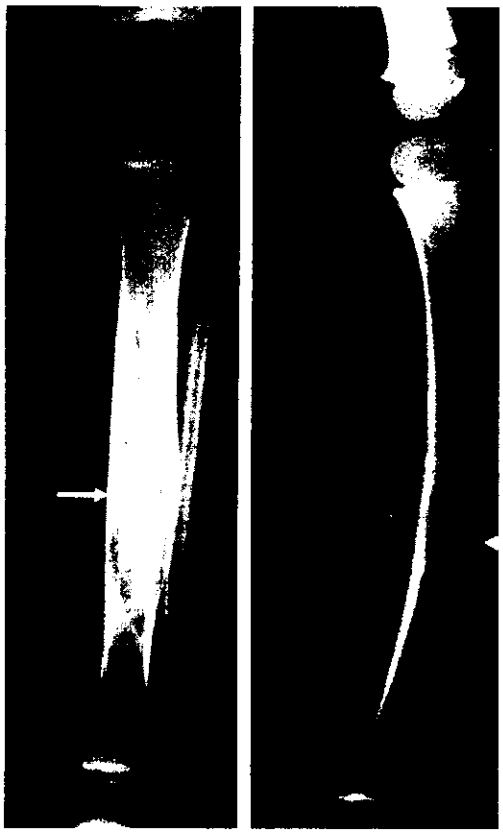


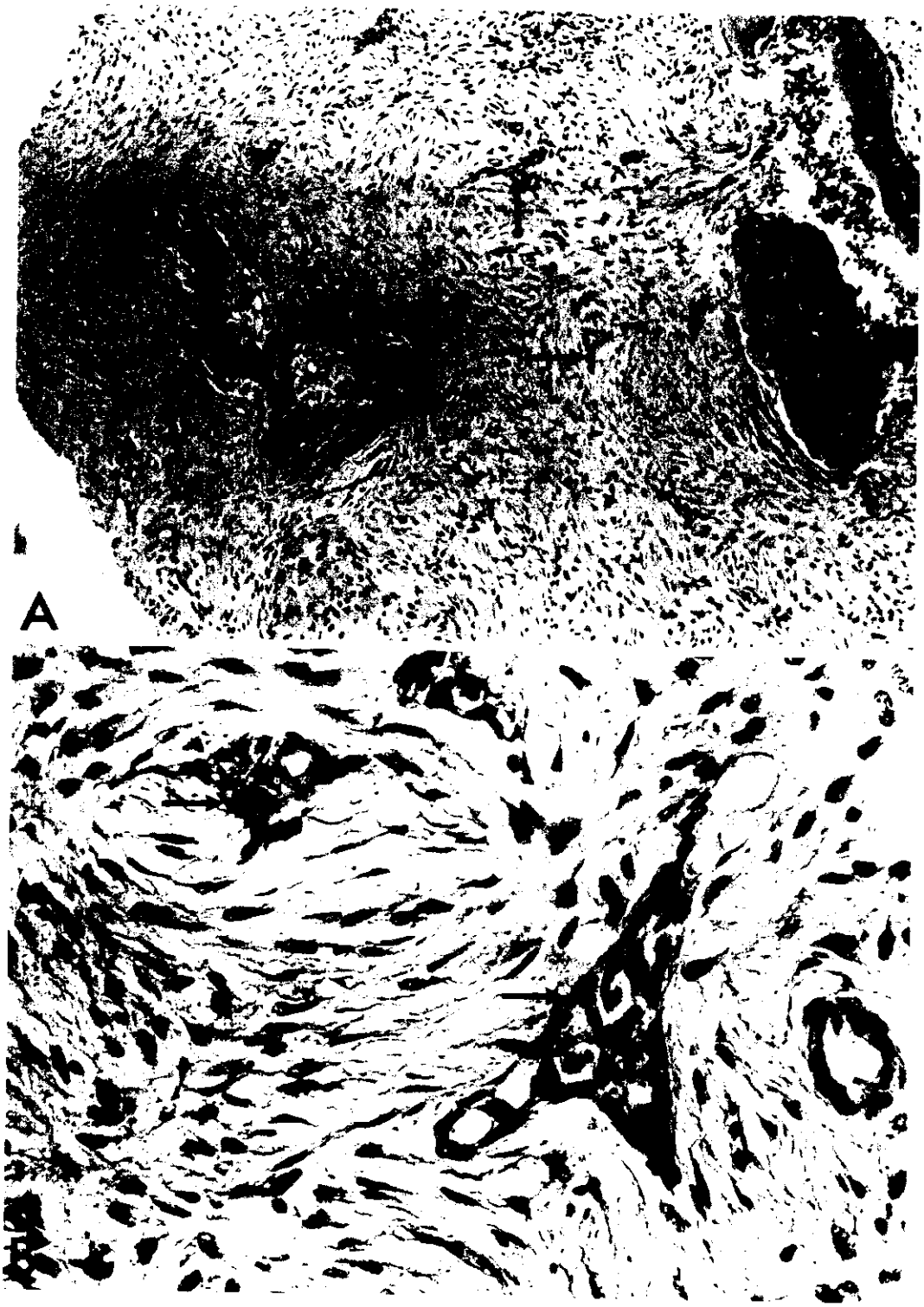
FIG. 5. Case 3. AP and lateral roentgenograms showing a large osteolytic multilocular lesion involving the midshaft of the tibia. The anterior cortex is thinned and slightly expanded with bowing of the tibia.

This benign process (osteofibrous dysplasia) is apparently of cortical origin and is defined by the histologic picture that shows the presence of woven bone trabeculae surrounded by rows of active osteoblasts, contrary to the classical fibrous dysplasia in which the osteoblasts on the surface of the bone trabeculae are usually flat and atrophic. This pattern indicates activity or aggressiveness of an otherwise typical fibrous dysplasia and does not justify the introduction of a new and confusing term[22] or the separation of these entities into two diseases. Osteofibrous dysplasia is not different from fibro-osseous dysplasia, which was used as a synonym for fibrous dysplasia by Jaffe.[11] Intracortical fibrous dysplasia, a term coined by Johnson,[12] is the preferred term by the authors who believe that the cortical location underlying the periosteum of a young growing child could easily explain the conspicuous osteoblastic activity of the process and may reflect the consequence of greater stress in lower leg bones.

In each of the three patients the roentgenographic features were characteristic of intracortical fibrous dysplasia. In Cases 2 and 3, the typical histologic features of osteofibrous dysplasia (ossifying fibroma) were found, *i.e.*, rims of active osteoblasts surrounded woven bone trabeculae within an abundant fibrous stroma that often has a storiform pattern around vessels and is rather loose. This pattern predominated and masked the scattered islands of epitheliallike cells. An erroneous diagnosis of fibrous or osteofibrous dysplasia was made by those who originally examined the histologic sections, and in one of the cases also by the consulted bone pathologist. Only after a careful study of all sections could the correct diagnosis of adamantinoma be established in the three cases.

Adamantinoma of the tibia is not as rare in children under ten years of age as has been reported in the literature;[11,16] it was found in two of 14 cases in our files (14.3%) and is probably often masked by the prominent fibrous dysplasialike component. That may explain the aggressive behavior of some cases of supposed fibrous (osteofibrous) dysplasia that have recurred in spite of repeated exci-

FIGS. 6A AND 6B. (A) Photomicrographs of Case 3 showing small scattered islands of epithelioid tumor cells (arrows) within a prominent fibrous stroma showing fibrous dysplasialike features (hematoxylin and eosin; original magnification, ×100). (B) The same tumor islands at a higher magnification (hematoxylin and eosin; original magnification ×400).



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3. Campanacci, M., C... and Gitelis, S.: Ad... The experience at t... Am. J. Surg. Patho...
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5. Changus, G. W., S... Malignant angiobl... adamantinoma of ...
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7. DeLarue, J., Cho... Adamantinome d... Ann. Anat. Pathol. ...
8. DeSantis, E., and F... ossa lunghe: Aspet... tinoma della tibia ... 28:129, 1977.
9. Eisenstein, W., an... of the tibia. An e... Lab. Med. 108:24(...
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REFERENCES

1. Albores-Saavedra, J., Diaz Gutierrez, D., and Altamirano Dimas, M.: Adamantinoma de la tibia. Observaciones ultraestructurales. Rev. Med. Hosp. Gen. Mexico City 31:241, 1968.
2. Baker, P. L., Dockerty, M. B., and Coventry, M. B.: Adamantinoma (so-called) of the long bones: Review of the literature and report of three new cases. J. Bone Joint Surg. 36A:704, 1954.
3. Campanacci, M., Giunti, A., Bertoni, F., Laus, M., and Gitelis, S.: Adamantinoma of the long bones: The experience at the Instituto Ortopedico Rizzoli. Am. J. Surg. Pathol. 5:533, 1981.
4. Campanacci, M., and Laus, M.: Osteofibrous dysplasia of the tibia and fibula. J. Bone Joint Surg. 63A:367, 1981.
5. Changus, G. W., Speed, J. S., and Stewart, F. W.: Malignant angioblastoma of bone: A reappraisal of adamantinoma of long bone. Cancer 20:540, 1957.
6. Cohen, D. M., Dahlin, D. C., and Pugh, D. G.: Fibrous dysplasia associated with adamantinoma of long bones. Cancer 15:515, 1962.
7. DeLarue, J., Chomette, G., and Brocheriou, C.: Adamantinome du tibia et "dysplasie fibreuse." Ann. Anat. Pathol. 9:373, 1964.
8. DeSantis, E., and Porfiri, B.: L'adamantinoma delle ossa lunghe: Aspetti ultrastrutturali di un adamantinoma della tibia. Arch. Putti Chir. Organi Mov. 28:129, 1977.
9. Eisenstein, W., and Pitcock, J. A.: Adamantinoma of the tibia. An eccrine carcinoma. Arch. Pathol. Lab. Med. 108:246, 1984.
10. Huvos, A. G., and Marcove, R. C.: Adamantinoma of long bones: A clinico-pathological study of fourteen cases with vascular origin suggested. J. Bone Joint Surg. 54A:148, 1975.
11. Jaffe, J. L.: Tumors and Tumorous Conditions of the Bones and Joints. Philadelphia, Lea & Febiger, 1958.
12. Johnson, L. C.: Congenital pseudoarthrosis, adamantinoma of long bone and intracortical fibrous dysplasia of the tibia. J. Bone Joint Surg. 54A:1355, 1972.
13. Kempson, R. L.: Ossifying fibroma of the long bones. Arch. Pathol. 82:218, 1966.
14. Llombart-Bosch, A., and Ortuno Pacheco, G.: Ultrastructural findings supporting the angioblastic nature of the so-called adamantinoma of the tibia. Histopathology 2:189, 1978.
15. Markel, S. F.: Ossifying fibroma of long bone: Its distinction from fibrous dysplasia and its association with adamantinoma of long bone. Am. J. Clin. Pathol. 69:91, 1978.
16. Moon, N. F., and Mori, H.: Adamantinoma of the appendicular skeleton—updated. Clin. Orthop. 204:215, 1986.
17. Mori, H., Yamamoto, S., Hiramatsu, K., Miura, I., and Moon, N. F.: Adamantinoma of the tibia: Ultrastructural and immunohistochemic study with reference to histogenesis. Clin. Orthop. 190:299, 1984.
18. Perez-Atayde, A. R., Kozakewich, H. P. W., and Vawter, G. F.: Adamantinoma of the tibia: An ultrastructural and immunohistochemical study. Cancer 55:1015, 1985.
19. Pieterse, A. S., Smith, P. S., and McClure, J.: Adamantinoma of long bones: Clinical, pathological and ultrastructural features. J. Clin. Pathol. 35:780, 1982.
20. Rosai, J.: Adamantinoma of the tibia: Electron microscopic evidence of its epithelial origin. Am. J. Clin. Pathol. 51:786, 1969.
21. Rosai, J., and Pinkus, G. S.: Immunohistochemical demonstration of epithelial differentiation in adamantinoma of the tibia. Am. J. Surg. Pathol. 6:427, 1982.
22. Schajowicz, F.: Tumors and Tumorlike Lesions of Bone and Joints. New York, Springer-Verlag, 1981.
23. Schajowicz, F., Cabrini, R. L., and Simes, R. J.: Microscopia electronica del "adamantinoma" de los huesos largos. Rev. Ortop. Traumatol. Lat. Am. 16:185, 1971.
24. Unni, K. K., Dahlin, D. C., Beabout, J. W., and Ivins, J. C.: Adamantinomas of long bones. Cancer 34:1976, 1974.
25. Weiss, S. W., and Dorfman, H. D.: Adamantinoma of long bone: An analysis of nine new cases with emphasis on metastasizing lesions and fibrous dysplasia-like changes. Hum. Pathol. 8:141, 1977.
26. Yoneyma, T., Winter, W. G., and Wilsow, L.: Tibial adamantinoma: Its histogenesis from ultrastructural studies. Cancer 40:1138, 1977.